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Case Report

Sudden death in adolescence caused by cardiac haemangioma

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ABSTRACT

Primary tumors of the heart in infants and children are rare. The types of heart tumors in pediatric age groups are generally different from those in adults. Cardiac myxoma is by far the most common tumor in adults, but in infants and adolescents the prevalent tumor of the heart is rhabdomyoma. Among benign cardiac tumors, cardiac hemangiomas are rare and often diagnosed post-mortem due to the lack of specific clinical symptoms and signs.

We report a case of sudden death due to cardiac hemangioma in an apparently healthy 15-year-old adolescent. The autopsy revealed a cardiac hemangioma located at the apex of the heart; the histopathological examination showed the tumor was a mixed capillary and arteriolar hemangioma, a very rare type of primary tumor in adolescents.

1. Introduction

Primary cardiac tumors in infants and children are rare. The types of tumors in pediatric age groups differ from those in adults. While cardiac myxomas are by far the most common primary tumors in adults, the most common primary tumor of the heart in infants and adolescents is rhabdomyoma.[1–5] Other types of primary heart tumors include cardiac hemangiomas, Purkinje cell tumors, cardiac teratomas, cardiac myxomas and malignant primary heart tumors. Among primary heart tumors, hemangiomas account for 5–10% of benign tumors.[3] Hemangiomas are usually asymptomatic and the clinical manifestations, if present, depend on tumor localization. Reports on symptomatic patients indicate arrhythmias and pericardial effusions, effort dyspnea, congestive heart failure, pseudoangina, outflow tract obstruction, and myocardial failure.[5–9]

Macroscopically, a cardiac hemangioma normally appears as a collection of larger and smaller vascular spaces. Microscopic variants consist in cavernous, capillary or arteriovenous hemangiomas.[3]

In this contribution, the authors report a representative case of sudden death due to cardiac hemangioma in an apparently healthy adolescent, and the pathology of the post-mortem findings.

2. Case summary

A 15-year-old boy died suddenly. His parents found him dead in his bed in the early morning hours. According to his parents, he had been living a normal life; there was no history of any significant medical illness, nor history of chest pain, dyspnea, arrhythmias or other symptoms of cardiac disease; no other young-age sudden death was reported in his family. Drug assumption was suspected. Legal authorities ordered an autopsy to investigate the cause of death.

An autopsy and external examination were performed. The external examination revealed a well-nourished adolescent, 175 cm in length and 65 Kg in weight. There were only a few linear, abraded wounds on the limbs. A complete autopsy was performed.

2.1. Heart

The pericardium was normal. The great vessels originating from the heart were anatomically regular. The heart weighed 240 g. Heart biometry was normal. Evidence of petechias on the epicardial surface; the endocardium was unremarkable. The coronary arteries originating from the right and left aortic sinuses were patent, with no evidence of atherosclerosis or other anomalies.

The macroscopic examination of the heart revealed a 3.0-cm intramural vascular neoformation located at the apex. The cut surface of the neoplasm was uniformly hemorrhagic (Fig. 1). All atrial and ventricular chambers appeared normal, except for the presence of the hemangioma in the apex of the heart. The left ventricular free wall measured 11.0 mm, the right ventricular free wall 4.0 mm, and the interventricular septum 15 mm.

2.2. Histological findings

The histological section of the cardiac tumor evidenced an irregular vascular structure. Massive thick-walled vascular venous

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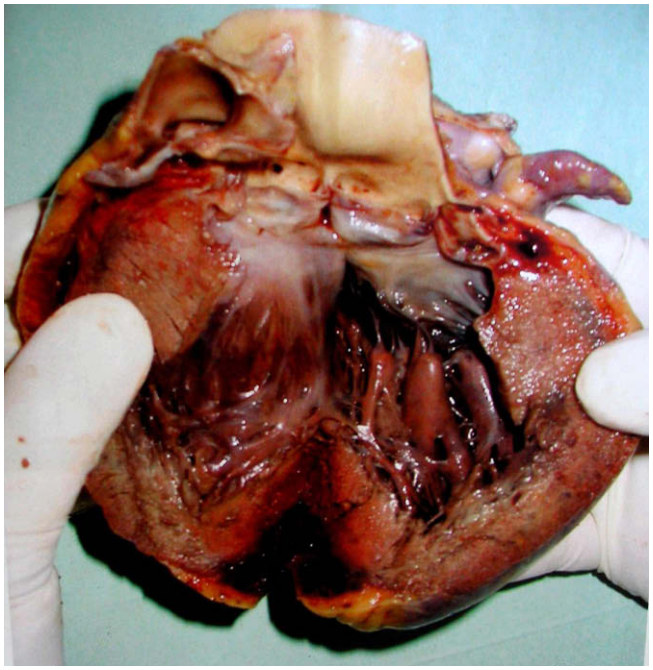

Fig. 1. Vascular neoformation, cm 3.0 in size, located at the apex.

spaces lined by flat endothelium replaced large portions of the posterior surface of both left and right ventricular walls and at the apex. There were also smaller thin-walled vessels and capillaries interposing the myocardial fibers, disrupting their normal course in many parts of both left and right ventricular walls (Figs. A–C). All toxicological analyses were negative.

2.3. Airways, gastrointestinal, brain and endocrine systems were unremarkable

3. Discussion and conclusion

Sudden death is an unexpected event, and related to cardiovascular diseases (especially coronary artery disease) and cardiomyopathies in more than 80%[13–15] of the cases, but a small percentage of these fatalities (approximately 0.0025%) stem from primary cardiac tumors such as cardiac hemangioma.[10–14]

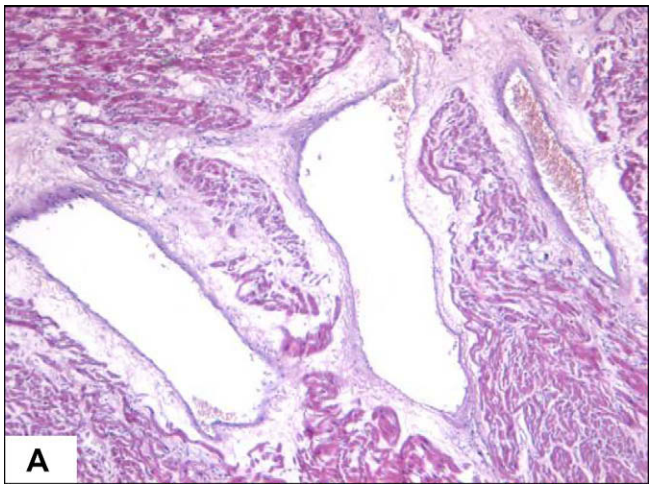

Fig. A. Cavernous vessels interrupting continuity of myocardial tissue (EE 40× and 100×).

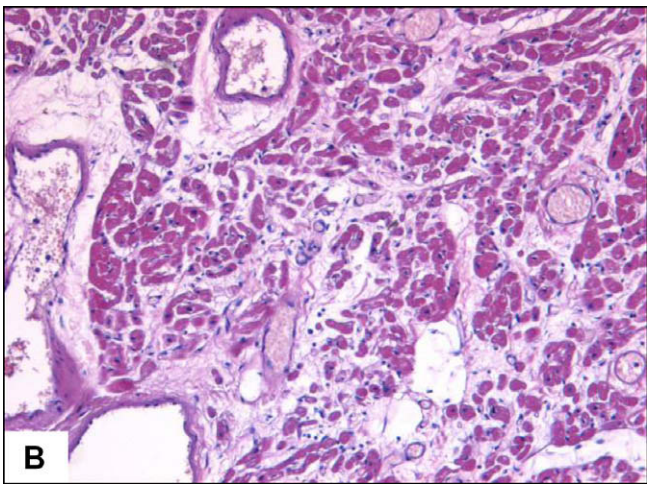

Fig. B. Capillary vessels mixed with cavernous component (EE 200×).

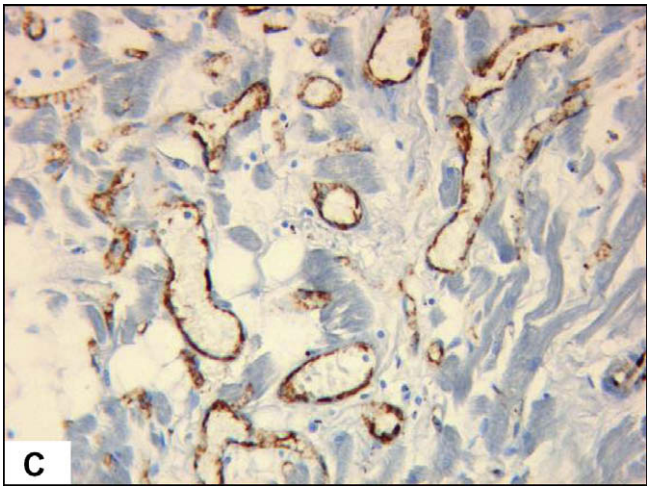

Fig. C. CD 34 endothelium immunostaining shows numerous capillary vessels (200×).

Cardiac hemangiomas are rare benign vascular tumors of the heart, most frequently located on the anterior wall of the right ventricle and less likely to be found on the lateral wall of the left ventricle.

Primary cardiac hemangiomas were first describe in 1983.[15] Few cases of sudden death due to cardiac hemangioma have been reported; in a review by McAllister, only 2.8% of 533 primary tumors and cysts of the heart and pericardium were hemangiomas.[6]

The most recent update from the Armed Forces Institute of Pathology (AFIP), encompassing 386 primary tumors of the heart collected between 1976 and 1993, reported only 55 tumors in infants and children younger than 16 years of age, 45 of which were benign and 11 considered malignant.[3]

The Cardiovascular Pathology Registry of the Academic Medical Center of Amsterdam (AMC) comprises 113 primary tumors of the heart and pericardium, 21 of which reported under the age of 16.[2] Only two cases of cardiac hemangiomas were reported in the AFIP[3] series and two cases by the AMC.[2]

Most affected patients are asymptomatic, and the cardiac hemangioma is often diagnosed incidentally at autopsy. Symptomatic

patients most commonly experience dyspnea, chest pain, right side heart failure, arrhythmias, pericarditis or pericardial effusion (which may be hemorrhagic), syncope and sudden death. The clinical presentation largely depends on the location and size of lesions. They may cause hemoperitoneum in case of epicardial location, and congestive heart failure or atrioventricular block when in an intramyocardial location.[16,17]

In this case, the boy showed no signs or symptoms of cardiac dysfunction prior to death, despite the location and size of the cardiac hemangioma (the autopsy showed the neoplasm had involved left and right ventricular walls) and the histopathological examination. There was no history of any sudden death in neither maternal nor paternal side of the family.

The disruption of the normal course of the myocardial fibers due to interposed neoformed vascular channels, observed microscopically, could give rise to fatal arrhythmias.

Cardiac hemangiomas are histologically classified into three types: tumors composed of multiple, dilated, thin-walled vessels (cavernous type), smaller capillary-like vessels (capillary type), and dysplastic malformed arteries and veins (arteriovenous type).[3] Cardiac hemangiomas often have combined features of capillary, cavernous, and arteriovenous hemangiomas. The cavernous and capillary types are reportedly encountered more frequently. In this case, the histological findings showed a mixed capillary and cavernous hemangioma.

Histologically, it is simple to diagnose a cardiac hemangioma. However, there are two types of tumors that should be considered in the differential diagnosis: myxomas and angiosarcomas. In particular, some low-grade angiosarcomas may be difficult to differentiate from hemangiomas, but the lack of mitotic activity, cellular pleomorphism, necrosis, and cellularity can distinguish a hemangioma from an angiosarcoma. So, without mitotic activity and nuclear pleomorphism, the diagnosis of cardiac hemangioma was formulated.

We believe the case reported is worth mentioning because a cardiac hemangioma is a rare cardiac tumor in an adolescent; considering the few cases reported in literature, it may contribute to the knowledge about cardiac sudden death.

Conflict of Interest

None declared.

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